Hydrocephalus Secondary to Hydrodynamic Disequilibrium in an Adult Patient with a Choroidal-Type Arteriovenous Malformation

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Summary

We describe an adult patient with an unruptured choroidal-type arteriovenous malformation (AVM) associated with progressive hydrocephalus. There was no evidence of mechanical obstruction of the ventricular system by the AVM nidus itself or a draining vein. However significant reflux into periventricular and transmedullary veins was demonstrated. Following partial targeted embolization of the AVM, no further reflux was observed, the patient's clinical deficits resolved, and the hydrocephalus improved. We suggest a hydrodynamic disorder as a potential pathomechanism of hydrocephalus in this adult patient with an unruptured AVM.

Introduction

Brain arteriovenous malformations (AVMs) are defined as abnormal collections of arteries and veins without an intervening capillary bed [1]. Typically, these lesions are discovered after an intracranial hemorrhage or new-onset seizure [2]. Neonate and infant, patients with unruptured AVMs, or more commonly, high-flow, pial arteriovenous or vein of Galen malformations, may present with hydrocephalus [3-5]. This presentation is exceedingly rare in adults harboring unruptured pial AVMs [6-11]. Geibprasert et al. have recently reported on eight patients (six adults and two children) with unruptured brain AVMs presenting with hydrocephalus [6]. In this series, the most common pathomechanism of hydrocephalus was mechanical obstruction of the ventricular pathways by an engorged draining vein or the AVM nidus itself. Only two patients (one adult and one child) with hydrocephalus presented without mechanical ventricular obstruction. The purported pathomechanism of hydrocephalus in these two patients was a hydrodynamic disequilibrium resulting from occluded dural sinuses that led to venous congestion and impaired absorption of cerebrospinal fluid (CSF) [12]. Here we describe an adult patient with an unruptured choroidal-type AVM with reflux into the periventricular and transmedullary veins. This patient presented with progressive hydrocephalus that improved after partial targeted embolization of the AVM and elimination of the venous reflux. This case provides further support for the hypothesis of a hydrodynamic disorder as a pathomechanism of hydrocephalus in adult patients with unruptured brain AVMs.

Case Report

Clinical Presentation

A 61-year-old man presented to medical attention with a five-month history of subjective gait instability. Otherwise, he was neurologically intact. An initial MRI brain showed hydrocephalus with evidence of mild transependymal edema (Figure 1A,B). Abnormal vascular flow

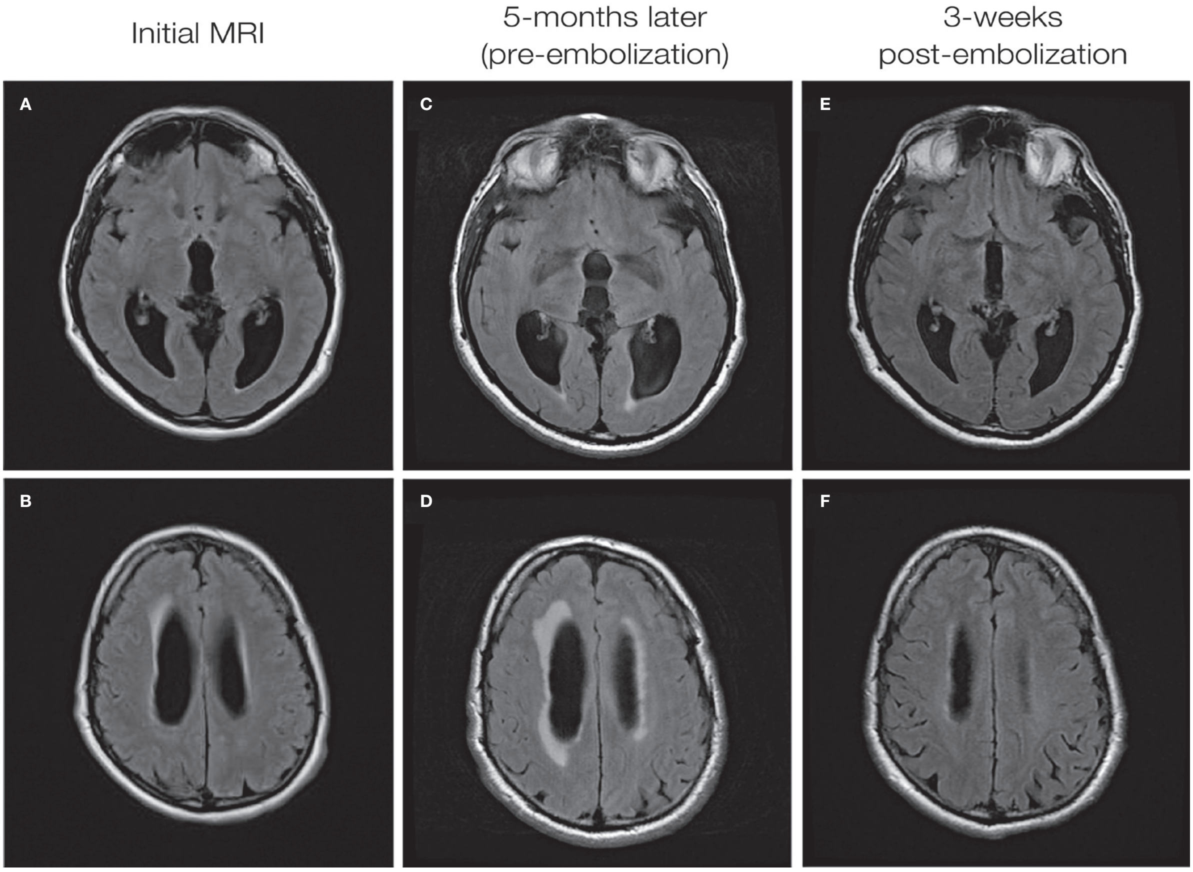

Figure 1 Axial FLAIR MRI performed at the time of presentation to medical attention (A and B), at the time of symptom progression (C and D), and 3-weeks (E and F) post-embolization. Note the progression of ventricular size and transependymal edema prior to treatment, and near resolution of hydrocephalus 3-weeks after partial targeted embolization of the choroidal-type AVM.

voids were noted in the region of the posterior third ventricle suggestive of an underlying vascular malformation (Figure 2A). Importantly, engorged periventricular and transmedullary veins were demonstrated, bilaterally (Figure 3). A 2-cm choroidal-type AVM was confirmed at conventional cerebral angiography. As suggested on the MRI, early venous reflux into distended periventricular and transmedullary veins was demonstrated (Figure 2B).

Over the next five months, the patient's gait instability worsened and was now accompanied by short-term memory and concentration problems. Repeat MRI brain demonstrated significant worsening of the hydrocephalus with significantly increased transependymal edema (Figure 1C,D). As previously demonstrated (Figure 2A), there was no evidence of mechanical obstruction of the ventricular system by a draining vein or the AVM nidus itself; however, the engorged periventricular and transmedullary veins appeared to be more prominent. Together, these findings suggested a hydrodynamic disorder as a putative pathomechanism of the patient's hydrocephalus secondary to venous congestion and impaired CSF reabsorption [12]. Given the patient's relatively rapid clinical deterioration, the decision was therefore made to proceed with partial targeted embolization of the AVM with the objective of reducing the degree of arteriovenous shunting and thereby ameliorating the venous hypertension, CSF malabsoprtion, and resultant hydrocephalus. The patient would then be referred for gamma knife radiosurgery as a definitive treatment for the AVM.

Intervention

Repeat catheter angiography again confirmed the choroidal-type AVM with prominent arterial feeders derived from the posterior choroidal arteries, bilaterally. A smaller, but important, arterial contribution was also derived from dural branches including (1) tentorial branches originating from the cavernous segment of the internal carotid arteries, bilaterally, and (2) middle meningeal artery branches originating from the external carotid arteries, bilaterally. Successful glue embolization (50% Histoacryl: 50% lipiodol) was performed of three vascular pedicles supplying the AVM nidus originating from the posterior cerebral arteries, bilaterally. Post-embolization control injections confirmed significantly reduced arteriovenous shunting with elimination of early reflux into the periventricular and transmedullary veins (Figure 2D). This was deemed a satisfactory result. Attempts at further embolization via the tentorial branches originating from the internal carotid arteries, bilaterally, proved unsuccessful as a result of significant proximal tortuosity and an inability to achieve a stable, distal microcatheter position.

Post-Embolization

Over the course of one week, the patient's neurocognitive decline was completely reversed and his gait instability significantly improved. An MRI brain performed three weeks post-embolization demonstrated a significant reduction in ventricular size with near complete resolution of transependymal edema (Figure 1E,F). The AVM nidus was also smaller in size consistent with reduced arteriovenous shunting (Figure 2C). Importantly, the previously demonstrated periventricular and transmedullary veins were no longer distended (Figure 2D). He is scheduled to undergo a second round of embolization to treat the hydrocephalus while awaiting definitive treatment for his AVM by gamma knife radiosurgery.

Discussion

Hydrocephalus in association with brain AVMs is most frequently secondary to intraventricular hemorrhage or subarachnoid hemorrhage with impaired CSF reabsorption [2]. In unruptured brain AVMs, however, associated hydrocephalus occurs by alternative mechanisms [6-11]. Geibprasert et al. recently reported on eight patients with unruptured AVMs and associated hydrocephalus [6]. The most common cause of hydrocephalus in the six adult patients was mechanical obstruction of the ventricular pathways by the AVM nidus itself or a deep draining vein. Alternatively, in two patients (one adult and one child), hydrocephalus resulted from a hydrodynamic disequilibrium. This hydrodynamic concept of hydrocephalus is based on the fact that CSF is absorbed not only via arachnoid granulations (Pacchionian bodies), as in the widely accepted bulk flow theory, but also via physiological transependymal flow at the level of the brain capillaries. Increased venous pressures in the periventricular and transmedullary veins, as a consequence of arteriovenous shunting towards periventricular and transmedullary veins in this patient, would therefore impede efficient CSF absorption by reducing the pressure gradient between the capillary bed and surrounding CSF spaces. This pathomechanism may help explain hydrocephalus and/or progressive dementia-like syndromes in adult patients with associated cranial dural arteriovenous shunts with cortical venous reflux [16-19]. It is already a well-described pathomechanism in the pediatric population; in particular, neonates and infants with vein of Galen AVMs or high-flow pial shunts [3-5]. In these very young, the arachnoid granulations are not yet fully matured, and CSF absorption is entirely dependent on a hydrodynamic mechanism. In some choroidal AVMs, increased CSF production may be a contributory factor, although this is decidedly rare, and difficult to substantiate [20]. Geibprasert et al. reported on a single adult patient who presented with hydrocephalus and an unruptured cerebellar AVM [6]. Mechanical ventricular obstruction was absent. Occlusion of the straight sinus was demonstrated with rerouting of venous drainage into cerebellar veins and severe posterior fossa congestion. The purported pathomechanism of hydrocephalus in this patient was hydrodynamic disequilibrium.

To the best of our knowledge, this is therefore only the second report to provide empirical evidence for hydrodynamic disequilibrium as a pathomechanism of hydrocephalus in an adult patient with an unruptured AVM. The practical relevance of this finding is that partial, targeted embolization of the AVM may represent a first-line treatment option for slowly progressive hydrocephalus in this patient population [6]. This statement is buttressed by the higher than usual complication rate observed with ventriculoperi-

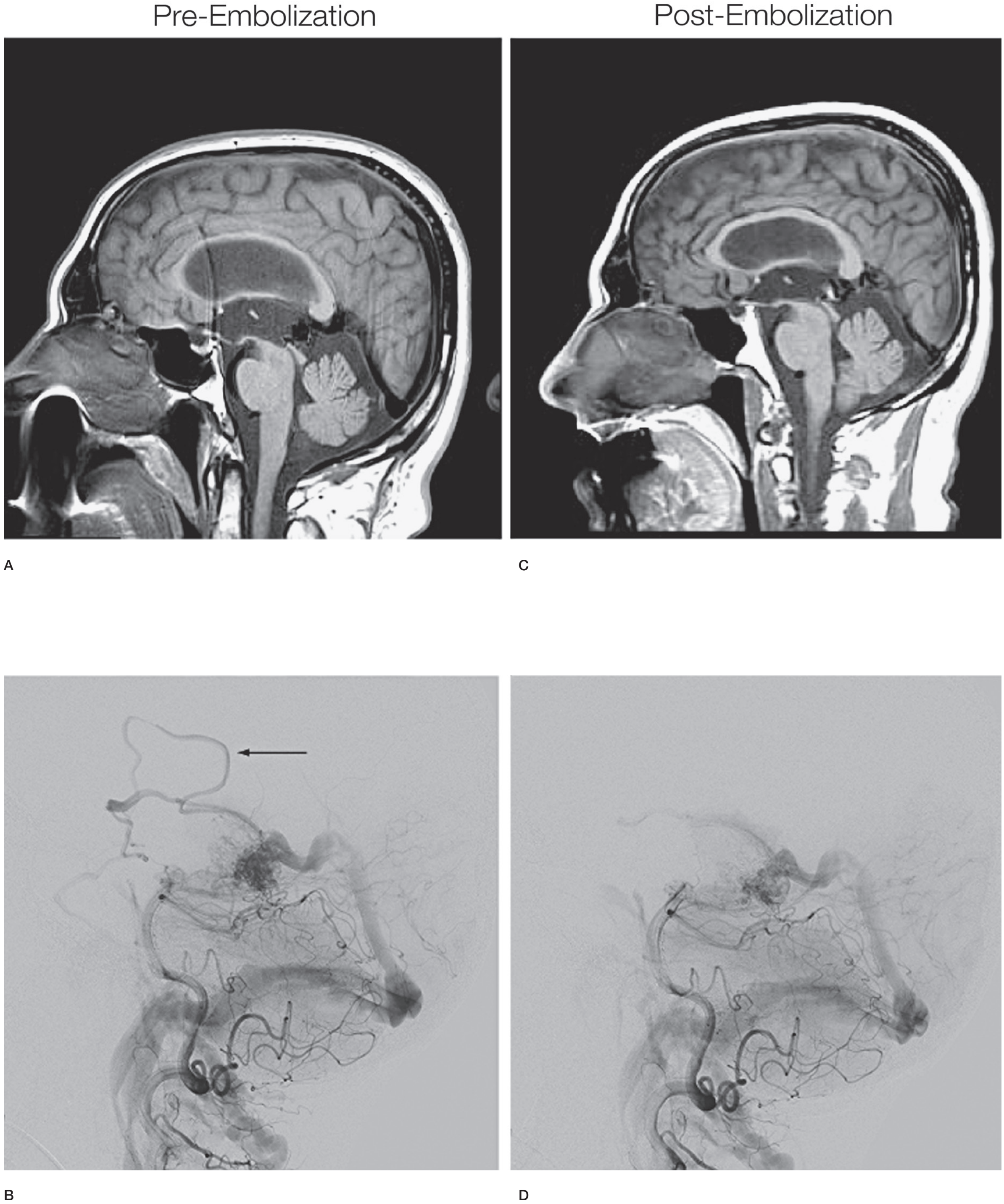

Figure 2 Sagittal T1-weighted MRI performed pre- (A) and post-embolization (C). On the pre-embolization image, note the choroidal-type AVM located in the region of the posterior third ventricle with associated hydrocephalus, i.e., ventricular enlargement and thinning of the corpus callosum. Importantly, the cerebral aqueduct is patent. On the post-embolization image, note the reduced arteriovenous shunting as evidenced by decreased flow voids in the vein of Galen and straight sinus, and corresponding improvement of the degree of hydrocephalus. Conventional digital subtraction angiogram performed pre- (B) and post-embolization (D). Note the absence of contrast filling of the transmedullary veins (arrow) post-embolization.

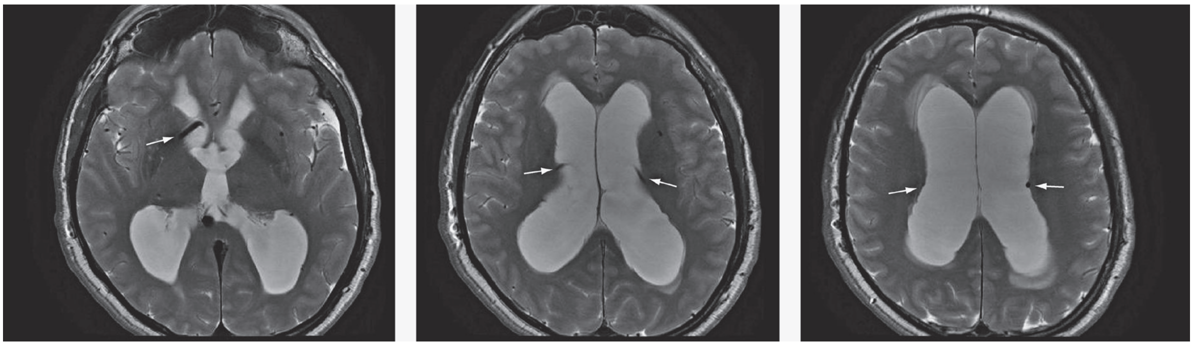

Figure 3 Axial T2-weighted MRI performed at the time of presentation to medical attention. Note the bilateral demonstration of engorged periventricular and transmedullary veins (arrows).

toneal shunting in patients with an underlying arteriovenous shunting lesion. Ventricular drainage, i.e., external ventricular drain insertion, ventriculoperitoneal shunting, or third ventriculostomy, should be reserved for refractory or life-threatening hydrocephalus.

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